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Laryngeal Neurofibroma Associated with Neurofibromatosis Type 2

Mutlu Cihangiroglu, Saim Yilmaz, Cahide Topsakal, Uzeyir Gok, Baki Altinsoy, and Bengu Cobanoglu

Summary: We present a case of laryngeal neurofibroma associated with neurofibromatosis type 2. Although laryngeal neurofibromas have previously been reported in cases of neurofibromatosis type 1, their presence has never been described in a patient with neurofibromatosis type 2.

Neurofibromatosis (NF) is an autosomal dominant disorder mainly characterized by abnormalities in the skin and nervous system. Although eight subtypes are known, NF1 and NF2 are the best described variants. NF1 is also known as *peripheral NF* with cardinal findings, including peripheral neurofibromas, café au lait spots, freckles, and Lisch nodules. NF2 is known as *central NF* and is generally characterized by bilateral vestibular schwannomas presenting with hearing loss during the second or third decades (1, 2). In addition to vestibular schwannomas, multiple meningiomas, spinal ependymomas, and nonvestibular schwannomas can also be seen in most cases (3–8).

Laryngeal nerve sheath tumors (neurinoma, neurofibroma) are very rare in patients with NF. They have mostly been reported as isolated cases (4). In the literature, laryngeal neurinomas have been reported to occur in two patients with NF1 and two patients with NF2 and neurofibromas have been reported to occur in 16 patients with NF1 (4–6, 8, 9). To the best of our knowledge, our case is the first to document a laryngeal neurofibroma in a patient with NF2. Another unique feature of our case is the coexistence of multiple intramedullary tumors, which has not previously been reported in a patient with a laryngeal neurofibroma.

Case Report

A 32-year-old woman presented with a history of cataract, hoarseness, and dysphonia since childhood, which had recently become worse. She was first examined at the Department of Ophthalmology, and surgery to treat the cataract was scheduled. Before the operation, consultation with an otorhinolaryngologist was requested to assess her relevant symptoms.

A physical examination revealed that the patient had weakness of the right arm and that three cutaneous lesions, each 1 to 2 cm in diameter, were located in the left arm and shoulder. The patient also had hearing disability for low frequencies. Laryngoscopy revealed a $2 \times 2 \times 3.5$ cm smooth surfaced submucosal supraglottic mass (Fig 1A). CT of the neck revealed the lesion to be a round and well-defined hypopharyngeal mass extended through and obliterating the left supraglottic space. It was hypoattenuated on unenhanced CT images and slightly enhanced with IV administration of contrast material (Fig 1B). The mass was heterogeneously hypointense on T1-weighted MR images (Fig 1C) and hyperintense on T2-weighted MR images (Fig 1D), with moderate homogenous enhancement after the administration of contrast material (Fig 1E). Incidentally, masses in both internal acoustic canals were noted at the time of investigation for laryngeal tumor; therefore, MR imaging studies of the brain and cervical spine were also performed. Bilateral vestibular schwannomas (Fig 1F) and multiple intramedullary masses (Fig 1G) (presumed to be ependymoma or astrocytoma) were delineated on these MR images. The patient was diagnosed as having NF2, and the laryngeal mass originating from the left aryepiglottic fold, which was suspected to be a neurofibroma or schwannoma, was totally resected. Histopathologic examination revealed the mass to be composed of multiple nodules of markedly vascular spindle cells and non-neoplastic connective tissue, which were consistent with neurofibroma (Fig 1H).

A month after the operation, the dysphonia had resolved almost completely. Further surgery for the vestibular schwannomas and cataract was planned. Because intramedullary tumors are asymptomatic, the patient was advised to undergo follow-up MR imaging of the spine at 1-year intervals.

Discussion

Diagnostic criteria for NF2 consists of bilateral eighth nerve masses or a first degree relative with NF2 and either unilateral eighth nerve mass or two of the following: neurofibroma, meningioma, glioma, schwannoma, or juvenile subcapsular lenticular opacity (1, 2). Because neural sheath tumors usually manifest after the second decade, skin lesions and cataracts are important in the early detection of children with NF2 (2). Surprisingly, however, in the present case, one of the initial findings was dysphonia caused by the laryngeal neurofibroma.

Laryngeal nerve sheath tumors (neurinoma, neurofibroma) have been rarely reported in association with NF. In the literature, a total number of 160 cases of laryngeal nerve tumors have been reported, and only 20 of them were associated with NF. In 16 of the 20 cases, laryngeal neurofibromas were documented in patients with NF1, and in four of the 20 cases, laryngeal schwannomas were detected, two in patients with NF1 and two in patients with NF2. To our knowledge, a laryngeal neurofibroma has never been de-

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From the Departments of Radiology (M.C., B.A.), Neurosurgery (C.T.), ENT (U.G.), and Pathology (B.C.), Firat University School of Medicine, Elazığ, and the Department of Radiology (S.Y.), Akdeniz University School of Medicine, Antalya, Turkey.

Address reprint requests to Mutlu Cihangiroglu, MD, Department of Radiology, Firat University School of Medicine, 23199 Elazığ, Turkey.



FIG 1. Images from the case of a 32-year-old woman who presented with progressive hoarseness and dysphonia.

A, Endoscopic examination shows an oval mass in the supraglottic area adjacent to the epiglottis (arrow).

B, Contrast-enhanced CT scan shows a slightly hypoattenuated mass (arrow) within the left supraglottic area extending to the left aryepiglottic fold.

C, Axial view T1-weighted (600/25 [TR/TE]) MR image reveals a hypointense mass (large arrow) with a slightly hyperintense periphery as compared with the center (small arrow).

D, Axial view T2-weighted (4000/117) MR image reveals a hyperintense mass (large arrow) with a slightly hypointense center as compared with the periphery (small arrow) in the same localization as that shown in B.

E, Axial view contrast-enhanced T1-weighted MR image shows diffuse enhancement of the tumor (arrow) within left supraglottic area.

F, Axial view contrast-enhanced T1-weighted image reveals densely enhanced bilateral vestibular schwannomas (arrows) at both cerebellopontine angles.

G, Sagittal view T1-weighted contrast-enhanced image confirms the intramedullary location of contrast-enhancing tumors (arrows) within the cervical and thoracic spine.

H, Histopathologic specimen from the laryngeal mass shows the tumor composed of neuronal fascicles and bundles within fibrous stroma, which is consistent with neurofibroma.

scribed as occurring in a patient with NF2, and our case is the first to report this association. Another interesting finding of our case was the presence of multiple intramedullar spinal masses. Although spinal cord tumors may be seen in patients with NF2, association of a laryngeal neurofibroma and spinal intramedullar tumors had not been reported to date.

Imaging findings of neurofibromas and neurinomas in locations other than the laryngeal nerve have been well described. On unenhanced CT scans, they are hypo- to slightly hyperattenuated; on T1-weighted MR images, they are iso- to hyperintense; and on T2-weighted MR images, they are mostly hyperintense. They almost always show a variable degree of enhancement on enhanced CT scans and MR images (1–3). CT and MR imaging findings of laryngeal nerve sheath tumors have been described in only a few cases. Plantet et al (4) reported two cases laryngeal schwannomas, with MR imaging findings in one and CT findings in both. On unenhanced CT scans, both schwannomas showed a hyperattenuated center with a peripheral area of low attenuation. MR imaging findings in one case included a slightly hyperintense center with peripheral hypointensity on T1-weighted images and a central heterogeneous intermediate signal intensity with peripheral hyperintensity on T2-weighted images. On the other hand, Martin et al (9) reported four cases of laryngeal neurofibroma that exhibited the similar marked T2 prolongation on MR images. In the present reported case, a central low attenuation area in the mass was shown on CT scans, and T2-weighted MR images also disclosed the slightly hypointense central areas. The central areas of T2 shortening could represent areas of fibrous tissue of laryngeal neurofibroma, as

seen in association with spinal plexiform neurofibromas. Although imaging findings in our cases were similar to those of the laryngeal neurofibromas reported by Stines et al (5), a differentiation between neurofibroma and schwannoma has not been attempted based on imaging findings, because the number of nerve sheath tumors reported with CT and MR imaging findings was very limited in this particular location.

In conclusion, dysphonia and hoarseness may be the only presenting symptoms, suggesting the possibility of a laryngeal nerve sheath tumor. Neurofibroma should be included in the differential diagnosis of laryngeal masses in patients with NF2.

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